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Xanthogranulomatous Pyelonephritis: A Retrospective Review of 21 Cases

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Abstract

Xanthogranulomatous Pyelonephritis is a relatively uncommon disorder of unknown etiology, a rare form of chronic pyelonephritis that is usually characterized by extensive destruction of the involved kidney. The renal parenchyma is replaced by lipid-laden macrophages referred to as xanthoma cells. It is increasingly being recognized as an important cause of renal morbidity worldwide. Patients who had nephrectomy had favorable outcome.

Keywords: Xanthogranulomatous Pyelonephritis; Xanthoma cells; Nephrectomy.

1. Introduction

Xanthogranulomatous Pyelonephritis is a rare and severe variant of chronic pyelonephritis typically characterized by diffuse destruction of the renal parenchyma and its replacement by inflammatory infiltrates of lipid-laden macrophages referred to as xanthoma cells. Most cases of Xanthogranulomatous Pyelonephritis are unilateral and often result in nonfunctioning kidney. They usually occur in association with nephrolithiasis, urinary tract obstruction and/or chronic urinary infection [1].

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The commonly associated organisms are Escherichia coli, Proteus mirabilis, Klebsiella, Pseudomonas aeruginosa and Staphylococcus aureus [1]

The disease starts from the pelvis and calyces and subsequently progresses to involve the entire renal parenchyma and adjacent tissues. Some have referred to this disease as the "great imitator" because clinically and radiologically, it is difficult to distinguish between it and other inflammatory diseases of the renal parenchyma and even renal cell carcinoma. We report a retrospective review of 21 cases managed at Urology and Nephrology Centre, Mansoura, Egypt from May 2001 to January 2007.

2. Materials and methods

A retrospective study was undertaken to review the cases of Xanthogranulomatous Pyelonephritis that presented at Urology and Nephrology Centre, Mansoura Egypt.

Result –A total number of 21 histology-proven cases from May 2001 to January 2007 treated at the above named institute were included in the study. There were 16 males (76.2%) and 5 females (23.8%) with M: F of 3.2:1. The age of patients at presentation ranges from 26 years to 75 years. All the patients presented with flank pain; 2 of the patients had nephrocutaneous fistula while one had paranephric abscess. Most of them had no growth of microorganisms on urine culture, 2 had elevated blood sugar. 13 patients (61.9%) had affectation of the left kidney while 8 patients (38.1%) had affectation of the right kidney. 76.2% had stone in the urinary tract on KUB X ray. All patients had nephrectomy done and recovered well with no significant postoperative complication.

Table 1

Sex	Percentage
Males	16 (76.2%)
Females	5 (23.8%)

Table 2

Kidney affected	Percentage
Left kidney	13 (61.9%)
Right kidney	8 (38.1%)

Table 3

Presenting symptom	Percentage
Flank pain	21 (100%)
Nephrocutaneous fistula	2 (9.5%)
Paranephric abcess	1(4.8%)

3. Discussion

Xanthogranulomatous Pyelonephritis is an uncommon disease condition and constitutes <1% of chronic pyelonephritis [2]. It was first described in detail by Schlagenhauter in 1916 [3].

The reported peak age incidence is 5th to 6th decade [4, 5]. However, it can occur at any age as it has been documented to have occurred in a 6 month old infant[6]. Our patients' age at presentation span through the 3rd to the 8th decade of life. It has a female to male ratio of 2:1[4, 5, 7]. This is at variance with findings from our study where we had more males with M: F of 3.2 to 1.0.

Symptoms are usually nonspecific with majority presenting with fever, flank pain, weight loss, malaise and anorexia. Pyuria is seen in 60 to 90% of patients. Common findings on examination include flank tenderness and palpable mass in the flank [8, 9]. From our study, all the patients had flank pain, 2 had nephrocutaneous fistula, 1 had paranephric abscess and 3 of the patients had microscopic haematuria. Haematuria has been known to be a rare feature of Xanthogranulomatous Pyelonephritis as previously reported[10]. Also contrary to previous report of its high prevalence in diabetics [10], only 2 of patients from this study had elevated blood sugar.

The primary factors implicated in the pathogenesis of Xanthogranulomatous Pyelonephritis include nephrolithiasis, urinary tract obstruction and chronic urinary infection [9, 11]. This correlates to a large extent with our findings where about 76.2% had stone in the urinary tract on the KUB x-ray.

Other possible interrelated factors include venous occlusion and haemorrhage, abnormal lipid metabolism, lymphatic blockage, failure of antimicrobial therapy in urinary tract infection, altered immunologic competence and renal ischemia [1, 12].

The organisms commonly implicated include Escherichia coli and Proteus mirabilis. Others include Pseudomonas aeruginosa, Klebsiella and Staphylococcus aureus. Majority of the patients in our study had no growth on urine culture.

There are 2 pathological forms of Xanthogranulomatous Pyelonephritis namely focal and diffuse forms. The diffuse form is characterised by generalised renal enlargement with multiple hypoechoic areas representing calyceal dilation and parenchymal destruction. The focal form has areas of hypoechoic mass which is often misdiagnosed as renal tumour [9, 11].

Radiological diagnosis of Xanthogranulomatous Pyelonephritis can be a challenge. However, computed tomography scan has been shown as one of the best investigations for preoperative evaluation, the diagnosis however can only be confirmed on histology. Radiological features which suggest diffuse Xanthogranulomatous Pyelonephritis includes renal enlargement, thickening of Gerota's fascia, perinephric fat strand and water density rounded areas in the renal parenchyma which represents dilated calyces and abscess cavities filled with pus and debris classically described as 'bear paw sign'. In the focal form, Computed Tomography usually shows a well-defined localized intra renal mass with fluid-like alteration [11, 13].

The differential diagnosis of Xanthogranulomatous Pyelonephritis includes renal cell carcinoma particularly the clear cell variant, wilms tumour, neuroblastoma and other causes of chronic pyelonephritis. The treatment of choice is antibiotics and partial or total nephrectomy [14]. All of our patients had nephrectomy and histology done. Histological features of Xanthogranulomatous Pyelonephritis on macroscopy shows enlarged kidney with thickened capsule, yellow nodules with or without areas of central necrosis. On microscopy, it shows yellow areas with multiple lipid laden macrophages otherwise called foam cells with extensive areas of inflammation and fibrosis [8, 9].

Sometimes the foam cells appear and are indeed misinterpreted as clear cells seen in renal cell carcinoma. This at times constitutes a diagnostic dilemma for the Histopathologist.

4. Conclusion

Xanthogranulomatous Pyelonephritis is a chronic and a relatively uncommon inflammatory condition involving the renal parenchyma. The definitive treatment is nephrectomy. However, to minimize morbidity and mortality, early identification and prompt treatment is important.

References

[1] C.-K. Chuang, M.-K. Lai, P.-L. Chang, M.-H. Huang, S.-H. Chu, C.-J. Wu, *et al.*, "Xanthogranulomatous pyelonephritis: experience in 36 cases," *The Journal of urology,* vol. 147, pp. 333-336, 1992.

[2] A. F. Leoni, P. Kinleiner, M. Revol, A. Zaya, and A. Odicio, "Xanthogranulomatous Pyelonephritis: review of 10 cases," *Arch Esp Urol,* vol. 62, pp. 259-271, 2009.

[3] F. Schlagenhaufer, "Uber eigentumliche Staphylomykosen der Nieren und des pararenalen Bindegewebes," *Frankfurt Z Pathol,* vol. 19, pp. 139-148, 1916.

[4] J. C. Gingell, J. Roylance, E. R. Davies, and J. B. Penry, "Xanthogranulomatous pyelonephritis," *Br J Radiol,* vol. 46, pp. 99-109, Feb 1973.

[5] U. S. Dwivedi, N. K. Goyal, V. Saxena, R. L. Acharya, S. Trivedi, P. B. Singh, *et al.*, "Xanthogranulomatous pyelonephritis: our experience with review of published reports," *ANZ journal of surgery,* vol. 76, pp. 1007-1009, 2006.

[6] S. Shinde, D. Kandpal, and S. Chowdhary, "Focal xanthogranulomatous pyelonephritis presenting as renal tumor," *Indian journal of nephrology,* vol. 23, p. 76, 2013.

[7] S. M. Goldman, D. S. Hartman, E. K. Fishman, J. P. Finizio, O. Gatewood, and S. S. Siegelman, "CT of xanthogranulomatous pyelonephritis: radiologic-pathologic correlation," *American journal of roentgenology,* vol. 142, pp. 963-969, 1984.

[8] R. J. Hendrickson, W. L. Lutfiyya, F. M. Karrer, P. D. Furness, 3rd, S. Mengshol, and D. D. Bensard, "Xanthogranulomatous pyelonephritis," *J Pediatr Surg*, vol. 41, pp. e15-7, Feb 2006.

[9] H. Cakmakci, N. Tasdelen, F. Obuz, E. Yilmaz, and A. Kovanlikaya, "Pediatric focal xanthogranulomatous pyelonephritis: dynamic contrast-enhanced MRI findings," *Clinical imaging*, vol. 26, pp. 183-186, 2002.

[10] S. Khalid, S. Zaheer, S. Zaheer, I. Ahmad, and M. Khalid, "Xanthogranulomatous pyelonephritis: Rare presentation of a rare disease," *South Asian journal of cancer*, vol. 2, p. 4, 2013.

[11] M. A. Al-Ghazo, I. F. Ghalayini, I. I. Matalka, N. S. Al-Kaisi, and Y. S. Khader, "Xanthogranulomatous pyelonephritis: analysis of 18 cases," *Asian Journal of Surgery*, vol. 29, pp. 257-261, 2006.

[12] M. Samuel, P. Duffy, S. Capps, P. Mouriquand, D. Williams, and P. Ransley, "Xanthogranulomatous pyelonephritis in childhood," *J Pediatr Surg*, vol. 36, pp. 598-601, Apr 2001.

[13] K. Shah, M. Parikh, P. Gharia, and P. R. Modi, "Xanthogranulomatous pyelonephritis-mimicking renal mass in 5-month-old child," *Urology*, vol. 79, pp. 1360-2, Jun 2012.

[14] R. Loffroy, B. Guiu, J. Watfa, F. Michel, J. Cercueil, and D. Krausé, "Xanthogranulomatous pyelonephritis in adults: clinical and radiological findings in diffuse and focal forms," *Clinical radiology*, vol. 62, pp. 884-890, 2007.